General

Guideline Title

Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis.

Bibliographic Source(s)

National Clinical Guideline Centre. Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. 32 p. (Clinical guideline; no. 163).

Guideline Status

This is the current release of the guideline.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- March 22, 2016 – Opioid pain medicines : The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.

Recommendations

Major Recommendations

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Note: The wording used in the recommendations in this guideline (for example, words such as 'offer' and 'consider') denotes the certainty with which the recommendation is made (the strength of the recommendations). See the end of the "Major Recommendations" field for further descriptions of the strength of recommendations.

Awareness of Clinical Features of Idiopathic Pulmonary Fibrosis

Be aware of idiopathic pulmonary fibrosis when assessing a patient with the clinical features listed below and when considering requesting a chest X-ray or referring to a specialist:

- Age over 45 years
- Persistent breathlessness on exertion
- Persistent cough
- Bilateral inspiratory crackles when listening to the chest
- Clubbing of the fingers
- Normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern

Diagnosis

Assess everyone with suspected idiopathic pulmonary fibrosis by:

- Taking a detailed history, carrying out a clinical examination (see recommendation above for clinical features) and performing blood tests to help exclude alternative diagnoses, including lung diseases associated with environmental and occupational exposure, with connective tissue diseases and with drugs and
- Performing lung function testing (spirometry and gas transfer) and
- Reviewing results of chest X-ray and
- Performing computed tomography (CT) of the thorax (including high-resolution images)

Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team (listed in Table 1 below), based on:

- The clinical features, lung function and radiological findings (see the recommendation in the section "Awareness of clinical features of idiopathic pulmonary fibrosis" above)
- Pathology when indicated (see the recommendation below)

At each stage of the diagnostic care pathway the multidisciplinary team should consist of a minimum of the healthcare professionals listed in Table 1, all of whom should have expertise in interstitial lung disease.

Table 1: Minimum Composition of Multidisciplinary Team Involved in Diagnosing Idiopathic Pulmonary Fibrosis

Stage of Diagnostic Care Pathway	Multidisciplinary Team Composition (All Healthcare Professionals Should Have Expertise in Interstitial Lung Disease)
After clinical evaluation, baseline lung function and computed tomography (CT)	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering performing bronchoalveolar lavage, and/or transbronchial biopsy or surgical lung biopsy Only some patients will have bronchoalveolar lavage or transbronchial biopsy but they may be being considered for surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy	Consultant respiratory physician Consultant radiologist

Stage of Diagnostic Care Pathway	Multidisciplinary Team Composition (All Healthcare Professionals Should Have Expertise in Interstitial Lung Disease) Consultant histopathologist Interstitial lung disease specialist nurse
	Multidisciplinary team coordinator
See Chapter 6.5 (Multidisciplinary Team) in the full guideline document for more information on the expertise of the multidisciplinary team.	

If a Confident Diagnosis Cannot Be Made

If the multidisciplinary team cannot make a confident diagnosis from clinical features, lung function and radiological findings, consider:

- Bronchoalveolar lavage or transbronchial biopsy and/or
- Surgical lung biopsy, with the agreement of the thoracic surgeon

Discuss with the person who may have idiopathic pulmonary fibrosis:

- The potential benefits of having a confident diagnosis compared with the uncertainty of not having a confident diagnosis and
- The increased likelihood of obtaining a confident diagnosis with surgical biopsy compared with bronchoalveolar lavage or transbronchial biopsy and
- The increased risks of surgical biopsy compared with bronchoalveolar lavage or transbronchial biopsy

When considering bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy take into account:

- The likely differential diagnoses and
- The person's clinical condition, including any comorbidities

If a confident diagnosis cannot be made continue to review the person under specialist care.

Information and Support

The consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with idiopathic pulmonary fibrosis, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management.

NICE has produced guidance on the components of good patient experience in adult NHS services. Follow the recommendations in Patient experience in adult NHS services (NICE clinical guideline 138).

An interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent.

Offer advice, support and treatment to aid smoking cessation to all people with idiopathic pulmonary fibrosis who also smoke, in line with the NICE guideline Smoking cessation services in primary care, pharmacies, local authorities and workplaces, particularly for manual working groups, pregnant women and hard to reach communities (NICE public health guidance 10).

Prognosis

Measure the initial rate of decline in the person's condition, which may predict subsequent prognosis, by using lung function test results (spirometry and gas transfer) at:

- Diagnosis and
- 6 months and 12 months after diagnosis. Repeat the lung function tests at shorter intervals if there is concern that the person's condition is deteriorating rapidly

Discuss prognosis with people with idiopathic pulmonary fibrosis in a sensitive manner and include information on:

- The severity of the person's disease and average life expectancy
- The varying courses of disease and range of survival
- Management options available

Do not use the 6-minute walk distance at diagnosis to estimate prognosis. (The 6-minute walk test may be useful for other purposes; see recommendation below.)

Management

Pulmonary Rehabilitation

Assess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6-minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality-of life assessment.

Repeat the assessment for pulmonary rehabilitation for people with idiopathic pulmonary fibrosis at 6-month or 12-month intervals.

If appropriate after each assessment, offer pulmonary rehabilitation including exercise and educational components tailored to the needs of people with idiopathic pulmonary fibrosis in general.

Pulmonary rehabilitation should be tailored to the individual needs of each person with idiopathic pulmonary fibrosis. Sessions should be held somewhere that is easy for people with idiopathic pulmonary fibrosis to get to and has good access for people with disabilities.

Best Supportive Care

Offer best supportive care to people with idiopathic pulmonary fibrosis from the point of diagnosis. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate:

- Information and support (see recommendation in the section "Information and support" above)
- Symptom relief
- Management of comorbidities
- Withdrawal of therapies suspected to be ineffective or causing harm
- End of life care

If the person is breathless on exertion consider assessment for:

- The causes of breathlessness and degree of hypoxia and
- Ambulatory oxygen therapy and long-term oxygen therapy and/or
- Pulmonary rehabilitation

If the person is breathless at rest consider:

- Assessment for the causes of breathlessness and degree of hypoxia and
- Assessment for additional ambulatory oxygen therapy and long-term oxygen therapy and
- The person's psychosocial needs and offering referral to relevant services such as palliative care services and
- Pharmacological symptom relief with benzodiazepines and/or opioids

Assess the oxygen needs of people who have been hospitalised with idiopathic pulmonary fibrosis before they are discharged.

If the person has a cough consider:

- Treatment for causes other than idiopathic pulmonary fibrosis (such as gastro-oesophageal reflux disease, post-nasal drip)
- Treating with opioids if the cough is debilitating
- Discussing treatment with thalidomide[1] with a consultant respiratory physician with expertise in interstitial lung disease if the cough is intractable.

Ensure people with idiopathic pulmonary fibrosis, and their families and carers, have access to the full range of services offered by palliative care teams. Ensure there is collaboration between the healthcare professionals involved in the person's care, community services and the palliative care team.

Disease-Modifying Pharmacological Interventions

There is no conclusive evidence to support the use of any drugs to increase the survival of people with idiopathic pulmonary fibrosis.

For guidance on pirfenidone for the management of idiopathic pulmonary fibrosis, refer to Pirfenidone for the treatment of idiopathic pulmonary fibrosis (NICE technology appraisal guidance 282).

Do not use any of the drugs below, either alone or in combination, to modify disease progression in idiopathic pulmonary fibrosis:

- Ambrisentan
- Azathioprine
- Bosentan
- Co-trimoxazole
- Mycophenolate mofetil
- Prednisolone
- Sildenafil
- Warfarin

Advise the person that oral N-acetylcysteine[2] is used for managing idiopathic pulmonary fibrosis, but its benefits are uncertain.

If people with idiopathic pulmonary fibrosis are already using prednisolone or azathioprine, discuss the potential risks and benefits of discontinuing, continuing or altering therapy.

Manage any comorbidities according to best practice. For gastro-oesophageal reflux disease, see Managing dyspepsia in adults in primary care (NICE clinical guideline 17).

Lung Transplantation

Discuss lung transplantation as a treatment option for people with idiopathic pulmonary fibrosis who do not have absolute contraindications. Discussions should:

- Take place between 3 and 6 months after diagnosis or sooner if clinically indicated
- Be supported by an interstitial lung disease specialist nurse
- Include the risks and benefits of lung transplantation
- Involve the person's family and carers with the person's consent
  (See the recommendations regarding best supportive care above.)

Refer people with idiopathic pulmonary fibrosis for lung transplantation assessment if they wish to explore lung transplantation and if there are no absolute contraindications. Ask the transplant centre for an initial response within 4 weeks.

Ventilation

A respiratory physician or specialist nurse with an interest in interstitial lung disease should discuss the poor outcomes associated with mechanical ventilation (including non-invasive mechanical ventilation) for respiratory failure with people with idiopathic pulmonary fibrosis. These discussions should ideally take place between 3 to 6 months after diagnosis or sooner if clinically indicated. (See recommendations about best supportive care above.)

Do not routinely offer mechanical ventilation (including non-invasive mechanical ventilation) to people with idiopathic pulmonary fibrosis who develop life-threatening respiratory failure.

Review and Follow-Up

In follow-up appointments for people with idiopathic pulmonary fibrosis:

- Assess lung function
- Assess for oxygen therapy
- Assess for pulmonary rehabilitation
- Offer smoking cessation advice, in line with the NGC summary of the NICE guideline Smoking cessation services in primary care, pharmacies, local authorities and workplaces, particularly for manual working groups, pregnant women and hard to reach communities (NICE public health guidance 10)
- Identify exacerbations and previous respiratory hospital admissions
- Consider referral for assessment for lung transplantation in people who do not have absolute contraindications (see recommendations above)
- Consider psychosocial needs and referral to relevant services as appropriate
- Consider referral to palliative care services
- Assess for comorbidities (which may include anxiety, bronchiectasis, depression, diabetes, dyspepsia, ischaemic heart disease, lung cancer

and pulmonary hypertension)

Consider follow-up of people with idiopathic pulmonary fibrosis:

- Every 3 months or sooner if they are showing rapid disease progression or rapid deterioration of symptoms or
- Every 6 months or sooner if they have steadily progressing disease or
- Initially every 6 months if they have stable disease and then annually if they have stable disease after 1 year

Footnotes

[1] At the time of publication (June 2013), thalidomide did not have a UK marketing authorisation for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Good practice in prescribing and managing medicines and devices for further information.

[2] At the time of publication (June 2013), N-acetylcysteine did not have a UK marketing authorisation. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Good practice in prescribing and managing medicines and devices for further information.

Definitions:

Strength of Recommendations

Some recommendations can be made with more certainty than others. The Guideline Development Group (GDG) makes a recommendation based on the trade-off between the benefits and harms of an intervention, taking into account the quality of the underpinning evidence. For some interventions, the GDG is confident that, given the information it has looked at, most patients would choose the intervention. The wording used in the recommendations in this guideline denotes the certainty with which the recommendation is made (the strength of the recommendation).

Interventions That Must (or Must Not) Be Used

The GDG usually uses 'must' or 'must not' only if there is a legal duty to apply the recommendation. Occasionally 'must' (or 'must not') is used if the consequences of not following the recommendation could be extremely serious or potentially life threatening.

Interventions That Should (or Should Not) Be Used – a 'Strong' Recommendation

The GDG uses 'offer' (and similar words such as 'refer' or 'advise') when confident that, for the vast majority of patients, an intervention will do more good than harm, and be cost-effective. Similar forms of words (for example, 'Do not offer…') are used when the GDG is confident that an intervention will not be of benefit for most patients.

Interventions That Could Be Used

The GDG uses 'consider' when confident that an intervention will do more good than harm for most patients, and be cost-effective, but other options may be similarly cost-effective. The choice of intervention, and whether or not to have the intervention at all, is more likely to depend on the patient's values and preferences than for a strong recommendation, and so the healthcare professional should spend more time considering and discussing the options with the patient.

Clinical Algorithm(s)

A NICE pathway on idiopathic pulmonary fibrosis is available at the National Institute for Health and Care Excellence (NICE) Web site .

Scope

Disease/Condition(s)

Idiopathic pulmonary fibrosis

Guideline Category

Diagnosis

Evaluation

Management

Rehabilitation

Risk Assessment

Treatment

Clinical Specialty

Allergy and Immunology

Critical Care

Family Practice

Internal Medicine

Pathology

Pulmonary Medicine

Radiology

Intended Users

Advanced Practice Nurses

Health Care Providers

Hospitals

Nurses

Patients

Physician Assistants

Physicians

Respiratory Care Practitioners

Guideline Objective(s)

To offer recommendations on the diagnosis and delivery of care to people with idiopathic pulmonary fibrosis, from initial suspicion of the disease, usually in primary care, through referral to a chest specialist, the role of multidisciplinary diagnostic and management teams and specific therapeutic interventions

Note: This guideline does not cover:

- Therapies for pulmonary hypertension as a complication of idiopathic pulmonary fibrosis
- Treatment of lung cancer as a complication of idiopathic pulmonary fibrosis
- Lung transplantation, other than timing and referral

Target Population

Adults (18 and older) with suspected or diagnosed idiopathic pulmonary fibrosis

Note: This guideline does not cover:

Children and young people (younger than 18)
People with a diagnosis of pulmonary fibrosis as a complication of:

- Connective tissue disorders (for example, systemic lupus erythematosus, rheumatoid arthritis, scleroderma, polymyositis and dermatomyositis)
- A known exogenous agent (for example, drug-induced disease or asbestosis)

Interventions and Practices Considered

Diagnosis/Evaluation/Prognosis

1. Awareness of clinical features of idiopathic pulmonary fibrosis
2. Detailed history, clinical examination, blood tests
3. Lung function testing (spirometry and gas transfer)
4. Chest X-ray
5. Computed tomography (CT) of thorax
6. Use of a multidisciplinary team for establishing diagnosis
7. Bronchoalveolar lavage and/or transbronchial biopsy
8. Surgical lung biopsy
9. Providing information and support to patient, families, carers
10. Predicting prognosis of disease

Treatment/Management

1. Use of 6-minute walk test for assessment for pulmonary rehabilitation
2. Pulmonary rehabilitation including exercise and educational components tailored to the individual
3. Best supportive care including ambulatory oxygen therapy, benzodiazepines, opioids, and thalidomide
4. Disease-modifying pharmacological interventions
    - Pirfenidone
    - N-acetylcysteine (benefits are uncertain)
    - The following drugs are not recommended: ambrisentan, azathioprine, bosentan, co-trimoxazole, mycophenolate mofetil, prednisolone, sildenafil, warfarin
5. Discussion of and referral for lung transplantation
6. Mechanical ventilation (not routinely recommended)
7. Review and follow-up

Major Outcomes Considered

- All-cause and idiopathic pulmonary fibrosis-related mortality
- Survival
- Change in predicted forced vital capacity
- Gas transfer: measurement of the carbon monoxide diffusing capacity of the lung
- Change in health-related quality of life measured using the Short Form-36 or Saint George's Respiratory Questionnaire and/or a measure of function such as the 6-minute walk test or EQ-5D
- Hospitalisations due to exacerbation of disease
- Adverse events
- Sensitivity and specificity of diagnostic tests
- Clinical and cost-effectiveness of interventions

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Developing the Review Questions and Outcomes

Review questions were developed in a PICO framework (patient, intervention, comparison and outcome) for intervention reviews, using population, presence or absence of factors under investigation (for example prognostic factors) and outcomes for prognostic reviews. This use of a framework guided the literature searching process, critical appraisal and synthesis of evidence, and facilitated the development of recommendations by the Guideline Development Group (GDG). The review questions were drafted by the NCGC technical team and refined and validated by the GDG. The GDG chose approximately 7 outcomes identifying which outcomes were critical to their decision making and which were important. This distinction helped the GDG to make judgements about the importance of the different outcomes and their impact on decision making. For example, mortality will usually be considered a critical outcome and would be given greater weight when considering the clinical effectiveness of an intervention than an important outcome with less serious consequences. The GDG decide on the relative importance in the review protocol before seeing the review.

Searching for Evidence

Clinical Literature Search

Systematic literature searches were undertaken to identify evidence within published literature in order to answer the review questions as per The Guidelines Manual 2012. Clinical databases were searched using relevant medical subject headings, free-text terms and study type filters where appropriate. Studies published in languages other than English were not reviewed. Where possible, searches were restricted to articles published in English language. All searches were conducted on core databases, MEDLINE, EMBASE and The Cochrane Library. The additional subject specific databases CINAHL and PsychINFO were used for some questions. All searches were updated on November 1, 2012. No papers published after this date were considered.

Search strategies were checked by looking at reference lists of relevant key papers, checking search strategies in other systematic reviews and asking the GDG for known studies. The questions, the study types applied, the databases searched and the years covered can be found in Appendices C and D of the full version of the guideline (see the "Availability of Companion Documents" field). During the scoping stage, a search was conducted for guidelines and reports on the websites listed below and on organisations relevant to the topic. Searching for grey literature or unpublished literature was not undertaken. All references sent by stakeholders were considered.

- Guidelines International Network database (www.g-i-n.net)
- National Guideline Clearinghouse (www.guideline.gov)
- National Institute for Health and Care Excellence (NICE) (www.nice.org.uk)
- National Institutes of Health Consensus Development Program (consensus.nih.gov)
- National Library for Health (www.library.nhs.uk)

Health Economic Literature Search

Systematic literature searches were also undertaken to identify health economic evidence within published literature relevant to the review questions. The evidence was identified by conducting a broad search relating to the guideline population in the National Health Service (NHS) Economic Evaluation Database (NHS EED), the Health Economic Evaluations Database (HEED) and health technology assessment (HTA) databases with no date restrictions. Additionally, the search was run on MEDLINE and EMBASE, with a specific economic filter, from 2010, to ensure recent publications that had not yet been indexed by these databases were identified. Studies published in languages other than English were

not reviewed. Where possible, searches were restricted to articles published in English language.

The search strategies for health economics are included in Appendix D of the full version of the guideline. All searches were updated on November 1, 2012. No papers published after this date were considered.

Evidence of Effectiveness

Clinical Evidence Inclusion/Exclusion Criteria

The inclusion and exclusion criteria were considered according to the PICO used in the protocols; see Appendix C of the full version of the guideline for full details. The GDG were consulted about any uncertainty regarding inclusion/exclusion of selected studies.

A major consideration in determining the inclusion and exclusion criteria in the protocol was the applicability of the evidence to the guideline population. The populations included in the review may differ for each review question, depending on the applicability of the data. See "Indirectness", section 3.3.8 in the full version of the guideline. The GDG acknowledged that data from interstitial lung disease (ILD) populations would include overlap between non-specific interstitial pneumonia (NSIP) and usual interstitial pneumonia (UIP), but agreed that the differences in clinical features and prognosis would not be a limitation for the evidence for diagnosis, pulmonary rehabilitation, best supportive care, psychosocial support and review and follow-up. However, a confirmed idiopathic pulmonary fibrosis population was specified for prognosis, pharmacological interventions, lung transplantation and ventilation. Idiopathic pulmonary fibrosis data from ILD populations was only included in these clinical areas if idiopathic pulmonary fibrosis alone was analysed separately.

Pre-1994 evidence was excluded by limiting searches to post 1994 data for review questions relating to diagnosis and prognosis only, as advances in computed tomography (CT) scanning have resulted in more consistent diagnosis of idiopathic pulmonary fibrosis after this time. No date restrictions were applied to any of the other clinical areas covered in this guideline.

Abstracts were included for three clinical areas; best supportive care, pulmonary rehabilitation and pharmacological interventions, on GDG advice due to the lack of evidence. Apart from those clinical areas abstracts were not included as evidence to inform other review questions, as the GDG considered that sufficient published evidence was available to inform decision making.

Health Economic Evidence Inclusion/Exclusion Criteria

Full economic evaluations (studies comparing costs and health consequences of alternative courses of action: cost–utility, cost-effectiveness, cost-benefit and cost-consequence analyses) and comparative costing studies that addressed the review question in the relevant population were considered potentially applicable as economic evidence.

Abstracts were assessed for applicability and included in the clinical review and for economic evidence for three clinical areas (best supportive care, pulmonary rehabilitation and pharmacological interventions). If assessed as potentially applicable, the authors were contacted for further information.

Studies were excluded which only reported cost per hospital (not per patient), or only reported the average cost effectiveness without disaggregated costs and effects. Posters, reviews, letters/editorials, foreign language publications and unpublished studies were also excluded. Studies judged to have an applicability rating of 'not applicable' were excluded (this included studies that took the perspective of non-organisation for economic co-operation and development countries).

Remaining studies were prioritised for inclusion based on their relative applicability to the development of this guideline and the study limitations. For example, if a high quality, directly applicable UK analysis was available other less relevant studies may not have been included. Where exclusions occurred on this basis, this is noted in the relevant section.

For more details about the assessment of applicability and methodological quality see the economic evaluation checklist (The Guidelines Manual, 2009) and the health economics research protocol in Appendix R of the full version of the guideline.

When no relevant economic analysis was found from the economic literature review, relevant UK NHS unit costs related to the compared interventions were presented to the GDG to inform the possible economic implication of the recommendation to make.

Number of Source Documents

The number of studies identified for each review question is provided in each review chapter of the full guideline document (see the "Availability of Companion Documents" field).

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Overall Quality of Outcome Evidence in GRADE (Grading of Recommendations Assessment, Development and Evaluation)

Level	Description
High	Further research is very unlikely to change confidence in the estimate of effect
Moderate	Further research is likely to have an important impact on confidence in the estimate of effect and may change the estimate
Low	Further research is very likely to have an important impact on confidence in the estimate of effect and is likely to change the estimate
Very Low	Any estimate of effect is very uncertain

Methods Used to Analyze the Evidence

Meta-Analysis of Randomized Controlled Trials

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

Note from the National Guideline Clearinghouse (NGC): This guideline was developed by the National Clinical Guideline Centre (NCGC) on behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Evidence of Clinical Effectiveness

Literature Review

The Research Fellows:

- Identified potentially relevant studies for each review question from the relevant search results by reviewing titles and abstracts – full papers were then obtained.
- Reviewed full papers against pre-specified inclusion/exclusion criteria to identify studies that addressed the review question in the appropriate population and reported on outcomes of interest (review protocols are included in Appendix C [see the "Availability of Companion Documents" field]). To minimise errors and any potential bias in the assessment, two reviewers independently assessed a random selection of studies. Any differences arising from this were then discussed with the Guideline Development Group (GDG).
- Critically appraised relevant studies using the appropriate checklist as specified in The Guidelines Manual.
- Extracted key information about the study's methods and results into evidence tables (evidence tables are included in Appendix F of the full version of the guideline).
- Generated summaries of the evidence by outcome (included in the relevant chapter write-ups) and produced evidence statements indicating the number of included studies, sample size (number randomised), direction of effect, uncertainty and Grading of Recommendations Assessment, Development and Evaluation (GRADE) quality rating:
    - Randomised studies: meta analysed, where appropriate and reported in GRADE profiles (for clinical studies) – see below for details
    - Observational studies: data presented as a range of values in adapted GRADE profiles
    - Diagnostic studies: data presented as a range of values in adapted GRADE profiles
    - Prognostic studies: data presented as a range of values in adapted GRADE profiles
    - Qualitative studies: each study summarised in a table where possible, otherwise presented in a narrative

Methods of Combining Clinical Studies

Data Synthesis for Intervention Reviews

Available Case Analysis

Estimates of effect from individual studies were based on available case analysis (ACA) where it was possible to extract these data. ACA was defined as analysis using all participants with data available for the outcome being considered. For example, for dichotomous outcomes, the denominator is the number of participants with available data and the numerator is the number who experienced the event. Participants for whom data for that outcome were not available are assumed to be missing at random. Where ACA was not possible data were reported as in the study and this is explained in the introduction of the relevant clinical review.

This method was used rather than intention-to-treat (ITT) analysis to avoid making assumptions about the participants for whom outcome data were not available, and rather assuming that those who drop out have the same event rate as those who continue. This also avoids incorrectly weighting studies in meta-analysis and overestimating the precision of the effect by using a denominator that does not reflect the true sample size with outcome data available.

ITT analysis is where all participants that were randomised are considered in the final analysis based on the intervention and control groups to which they were originally assigned. It was assumed that participants in the trials lost to follow-up did not experience the outcome of interest (categorical outcomes) and they would not considerably change the average scores of their assigned groups (for continuous outcomes). It is important to note that ITT analyses tend to bias the results towards no difference. ITT analysis is a conservative approach to analyse the data, and therefore the effect may be smaller than in reality.

Meta-Analyses

Where possible, meta-analyses were conducted to combine the results of studies for each review question using Cochrane Review Manager (RevMan5) software. Fixed-effects (Mantel-Haenszel) techniques were used to calculate pooled risk ratios (relative risk) for the binary outcomes. The continuous outcomes were analysed using an inverse variance method for pooling weighted mean differences and where the studies had different scales, standardised mean differences were used.

Statistical heterogeneity was assessed by considering the chi-squared test for significance at $p < 0.1$ or an I-squared inconsistency statistic of >50% to indicate significant heterogeneity. Assessments of potential differences in effect between subgroups were based on the chi-squared tests for heterogeneity statistics between subgroups.

The means and standard deviations of continuous outcomes were required for meta-analysis. However, in cases where standard deviations were not reported, the standard error was calculated if the p-values or 95% confidence intervals were reported and meta-analysis was undertaken with the mean and standard error using the generic inverse variance method in Cochrane Review Manager (RevMan5) software. When the only evidence was based on studies which only presented means, this information was summarised in the GRADE tables without calculating the relative and absolute effect.

For binary outcomes, absolute event rates were also calculated using the GRADEpro software using event rate in the control arm of the pooled results.

Data Synthesis for Prognostic Factor Reviews

Odds ratio, relative risks or hazard ratios, with their 95% confidence intervals, from multivariate analyses were extracted from the papers, and standard errors were calculated from the 95% confidence intervals. The log of the effect size with its standard error was entered into the generic inverse variance technique in the Cochrane Review Manager (RevMan5) software (http://ims.cochrane.org/revman). Studies were not combined in a meta-analysis for observational studies.

The quality of studies was assessed and presented in an adapted GRADE profile according to criteria stated in the methodology checklist for prognostic studies in the guidelines manual. Results were reported as ranges.

Data Synthesis for Diagnostic Test Accuracy Review

Evidence for diagnostic data was evaluated by study, using version two of the Quality Assessment of Diagnostic Accuracy Studies checklists (QUADAS-2) (http://www.bris.ac.uk/quadas/quadas-2). For diagnostic test accuracy studies, the following outcomes were reported: sensitivity, specificity, positive predictive value and negative predictive value. In cases where the outcomes were not reported, 2 by 2 tables were constructed from raw data to allow calculation of these accuracy measures. Summary receiver operative characteristic (ROC) curves, would have been generated if appropriate; however, there were no data in the diagnostic reviews included in this guideline that could be

combined to produce an ROC curve or diagnostic meta-analysis.

Data Synthesis for Qualitative Review

Themes were identified from these studies by two reviewers independently, and then verified jointly. These themes were supplemented with data from surveys where available. Common themes relevant to the question are reported in a narrative in the guideline text.

Appraising the Quality of Evidence by Outcomes

The evidence for outcomes from the included randomized control trials (RCT) and observational studies were evaluated and presented using the 'Grading of Recommendations Assessment, Development and Evaluation (GRADE) toolbox' developed by the international GRADE working group (http://www.gradeworkinggroup.org/). The software (GRADEpro) developed by the GRADE working group was used to assess the quality of each outcome, taking into account individual study quality and the meta-analysis results. The summary of findings was presented as two separate tables in this guideline. The 'Clinical/Economic Study Characteristics' table includes details of the quality assessment while the 'Clinical/Economic Summary of Findings' table includes pooled outcome data, where appropriate, an absolute measure of intervention effect and the summary of quality of evidence for that outcome. In this table, the columns for intervention and control indicate the sum of the sample size for continuous outcomes. For binary outcomes such as number of people with an adverse event, the event rates (n/N: number of people with events divided by sum of number of people) are shown with percentages. Reporting or publication bias was only taken into consideration in the quality assessment and included in the Clinical Study Characteristics table if it was apparent.

Each outcome was examined separately for the quality elements listed and defined in Table 2 and each graded using the quality levels listed in Table 3 (see the full version of the guideline). The main criteria considered in the rating of these elements are also discussed in the full version of the guideline. Footnotes were used to describe reasons for grading a quality element as having serious or very serious problems. The ratings for each component were summed to obtain an overall assessment for each outcome.

Evidence of Cost-Effectiveness

Literature Review

The Health Economist:

- Identified potentially relevant studies for each review question from the economic search results by reviewing titles and abstracts – full papers were then obtained.
- Reviewed full papers against pre-specified inclusion/exclusion criteria to identify relevant studies (see below for details).
- Critically appraised relevant studies using the economic evaluations checklist as specified in The Guidelines Manual (see the "Availability of Companion Documents" field).
- Extracted key information about the study's methods and results into evidence tables (evidence tables are included in Appendix F in the full version of the guideline).
- Generated summaries of the evidence in NICE economic evidence profiles (included in the relevant chapter write-ups) – see below for details.

NICE Economic Evidence Profiles

The NICE economic evidence profile has been used to summarise cost and cost-effectiveness estimates. The economic evidence profile shows, for each economic study, an assessment of applicability and methodological quality, with footnotes indicating the reasons for the assessment. These assessments were made by the health economist using the economic evaluation checklist from The Guidelines Manual. It also shows incremental costs, incremental outcomes (for example, quality-adjusted life-years [QALYs]) and the incremental cost-effectiveness ratio from the primary analysis, as well as information about the assessment of uncertainty in the analysis. See Table 6 in the full version of the guideline for more details.

If a non-UK study was included in the profile, the results were converted into pounds sterling using the appropriate purchasing power parity.

Where economic studies compare multiple strategies, results are not reported in the standard economic profile but are instead presented at the end of the relevant chapter in an alternative table. The study is summarised as a whole in a descriptive manner.

Undertaking New Health Economic Analysis

As well as reviewing the published economic literature for each review question, as described above, new economic analyses were undertaken by the Health Economist in priority areas. Priority areas for new health economic analysis were agreed by the GDG after formation of the review questions and consideration of the available health economic evidence. Additional data for the analyses were identified as required through additional literature searches undertaken by the Health Economist, and discussion with the GDG. Model structure, inputs and assumptions were

explained to and agreed by the GDG members during meetings, and they commented on subsequent revisions.

See Appendices G, J, K, and L in the full version of the guideline for economic evidence tables as well as details of the health economic analyses undertaken for the guideline.

Methods Used to Formulate the Recommendations

Expert Consensus

Informal Consensus

Description of Methods Used to Formulate the Recommendations

Note from the National Guideline Clearinghouse (NGC)" This guideline was developed by the National Clinical Guideline Centre (NCGC) on behalf of the National Institute for Health and Care Excellence (NICE). See the "Availability of Companion Documents" field for the full version of this guidance.

Guideline Development Group (GDG)

A multidisciplinary Guideline Development Group (GDG) comprising professional group members and consumer representatives of the main stakeholders developed this guideline (see the section on Guideline Development Group Membership and acknowledgements in the full version of the guideline).

The group met every 4-5 weeks during the development of the guideline.

Staff from the NCGC provided methodological support and guidance for the development process. The team working on the guideline included a project manager, systematic reviewers, health economists and information scientists. They undertook systematic searches of the literature, appraised the evidence, conducted meta-analysis and cost effectiveness analysis where appropriate and drafted the guideline in collaboration with the GDG.

Developing Recommendations

Over the course of the guideline development process, the GDG were presented with:

- Evidence tables of the clinical and economic evidence reviewed from the literature. All evidence tables are in Appendix F and G.*
- Summary of clinical and economic evidence and quality (as presented in Chapters 5 to 13).
- Forest plots (Appendix E).*
- A description of the methods and results of the cost-effectiveness analysis undertaken for the guideline (Appendix L).*

* See the full version of the guideline for appendices and specific chapters (see the "Availability of Companion Documents" field).

Recommendations were drafted on the basis of the GDG interpretation of the available evidence, taking into account the balance of benefits and harms, quality of evidence, and costs. When clinical and economic evidence was of poor quality, conflicting or absent, the GDG drafted recommendations based on consensus. Expert advisors were invited to provide advice on how to interpret the identified evidence. The considerations for making consensus based recommendations included the balance between potential harms and benefits, economic implications compared to the benefits, current practices, recommendations made in other relevant guidelines, patient preferences and equality issues. The consensus recommendations were done through discussions in the GDG, or methods of formal consensus were applied. The GDG considered whether the uncertainty was sufficient to justify delaying making a recommendation to await further research, taking into account the potential harm of failing to make a clear recommendation.

The main considerations specific to each recommendation are outlined in the Evidence to Recommendation Sections preceding the recommendation section in each chapter.

The guideline recommends some drugs for indications for which they do not have a UK marketing authorisation at the date of publication, if there is evidence to support that use.

Rating Scheme for the Strength of the Recommendations

Strength of Recommendations

Some recommendations can be made with more certainty than others. The Guideline Development Group (GDG) makes a recommendation based on the trade-off between the benefits and harms of an intervention, taking into account the quality of the underpinning evidence. For some interventions, the GDG is confident that, given the information it has looked at, most patients would choose the intervention. The wording used in the recommendations in this guideline denotes the certainty with which the recommendation is made (the strength of the recommendation).

Interventions That Must (or Must Not) Be Used

The GDG usually uses 'must' or 'must not' only if there is a legal duty to apply the recommendation. Occasionally 'must' (or 'must not') is used if the consequences of not following the recommendation could be extremely serious or potentially life threatening.

Interventions That Should (or Should Not) Be Used – a 'Strong' Recommendation

The GDG uses 'offer' (and similar words such as 'refer' or 'advise') when confident that, for the vast majority of patients, an intervention will do more good than harm, and be cost-effective. Similar forms of words (for example, 'Do not offer…') are used when the GDG is confident that an intervention will not be of benefit for most patients.

Interventions That Could Be Used

The GDG uses 'consider' when confident that an intervention will do more good than harm for most patients, and be cost-effective, but other options may be similarly cost-effective. The choice of intervention, and whether or not to have the intervention at all, is more likely to depend on the patient's values and preferences than for a strong recommendation, and so the healthcare professional should spend more time considering and discussing the options with the patient.

Cost Analysis

In general, an intervention was considered to be cost-effective if either of the following criteria applied (given that the estimate was considered plausible):

a. The intervention dominated other relevant strategies (that is, it was both less costly in terms of resource use and more clinically effective compared with all the other relevant alternative strategies), or
b. The intervention cost less than £20,000 per quality-adjusted life-year (QALY) gained compared with the next best strategy.

If the Guideline Development Group (GDG) recommended an intervention that was estimated to cost more than £20,000 per QALY gained, or did not recommend one that was estimated to cost less than £20,000 per QALY gained, the reasons for this decision are discussed explicitly in the 'from evidence to recommendations' section of the relevant chapter with reference to issues regarding the plausibility of the estimate or to the factors set out in the National Institute for Health and Care Excellence (NICE) report 'Social value judgements: principles for the development of NICE guidance'.

If a study reported the cost per life year gained but not QALYs, the cost per QALY gained was estimated by multiplying by an appropriate utility estimate to aid interpretation. The estimated cost per QALY gained is reported in the economic evidence profile with a footnote detailing the life-years gained and the utility value used. When QALYs or life years gained are not used in the analysis, results are difficult to interpret unless one strategy dominates the others with respect to every relevant health outcome and cost.

Detailed economic evidence is reviewed for each review question in the full version of the guideline. In addition the following are provided:

- Appendix G: Economic Evidence Tables
- Appendix J: Costing of a Multidisciplinary Team (MDT) in the context of an interstitial lung disease (ILD) network: Finding the incremental cost of involving an MDT in the idiopathic pulmonary fibrosis diagnostic pathway
- Appendix K: Placing the diagnostic clinical evidence into an economic framework for decision making
- Appendix L: Cost-effectiveness analysis – pulmonary rehabilitation for patients with idiopathic pulmonary fibrosis

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation

Validation Process

The guidance is subject to a six week public consultation and feedback as part of the quality assurance and peer review the document. All comments received from registered stakeholders were responded to in turn and posted on the National Institute for Health and Care Excellence (NICE) Web site.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of evidence supporting the recommendations is not specifically stated.

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- Improved diagnosis, management and provision of supportive care for patients with suspected or known idiopathic pulmonary fibrosis.
- Improved patient counseling.

See the "Trade-off between clinical benefits and harms" sections of the full version of the original guideline document for additional details about benefits of specific interventions.

Potential Harms

Adverse Effects of Invasive Procedures and Pharmacological Interventions

- The guideline development group (GDG) acknowledged the increased risk of adverse events associated with biopsy (bronchoalveolar lavage, transbronchial biopsy and surgical lung biopsy). No direct harms to a person suspected of idiopathic pulmonary fibrosis was attributed to multidisciplinary team (MDT) discussions, but the GDG did recognise that harms associated with an incorrect diagnosis made by an MDT and subsequent inappropriate management plan may result in reduced quality of life. However, the GDG felt that the diagnostic accuracy and precision is dependent on the expertise of the clinicians, radiologists and histopathologists. The GDG acknowledged that typically a thoracic surgeon would be present at an MDT to aid surgical planning and further reduce the risk of an incorrect diagnosis being made.
- The GDG considered the incremental benefit of conducting a surgical biopsy against biopsy sampling error and adverse events such as risk of infection, haemoptysis and pneumothorax. In a proportion of the people with possible idiopathic pulmonary fibrosis, the risks of performing a surgical lung biopsy outweigh the benefits of confirming diagnosis.
- The GDG discussed the small risks associated with 6-minute walk test ('exertional tests'), such as fainting, but did not consider that people with idiopathic pulmonary fibrosis undergoing the 6-minute walk test would be pushed to this extreme.
- The GDG considered exercise exertion beyond a person's normal capacity to be the only harm associated with pulmonary rehabilitation, but that this risk was unlikely due to the programmes being conducted by trained health professionals (physiotherapists and nurses).
- The potential harms of oxygen therapy are uncertain. Ambulatory oxygen therapy requires the patient to carry portable oxygen, and the benefits of the oxygen need to be balanced against the extra weight being carried. People with idiopathic pulmonary fibrosis may feel inhibited about using ambulatory oxygen, which is easily visible, in public places.
- It was recognised that there may be harms associated with inappropriate oxygen management if patients do not get followed-up in a timely manner and if there has been a change in symptoms due to disease progression or acute exacerbation. Currently, there may be a minority of patients who are discharged home and have oxygen requirements reassessed when they are back in the community, which carries a risk that for these patients oxygen management may not be optimal. Reassessing oxygen requirements prior to discharge brings no appreciable harm and will allow for optimal oxygen management following a potential change in the clinical status of the patient, thereby bringing clinical benefit and reducing the risk of clinical harm.

- The precautions for thalidomide use were acknowledged as were the uncertainties regarding the long term harms of thalidomide for cough, which are unknown.
- There was the recognition that patients can demonstrate serious adverse events profiles with the pharmacological interventions and may require withdrawal of ineffective therapies and thus may need the expertise of the interstitial lung disease (ILD) team to tailor alternative regimens for patients. Therefore, the GDG recognised the importance of ILD teams remaining involved in a patient's care even once they have been referred to the palliative care teams.
- As well as considering a patient's prognosis and clinical suitability for lung transplantation, the GDG acknowledged that a patient's social, financial and mental well-being (support from family and carers, and psychosocial support) would have a considerable impact on their eligibility for an invasive procedure. The GDG agreed that a patient should also be assessed on their social and mental capacity for lung transplantation. Complications associated with transplantation may include cellular or humeral rejection, infection, and primary organ dysfunction and airway complications.

See the "Trade-off between clinical benefits and harms" sections of the full version of the original guideline document for additional details about harms of specific interventions.

Qualifying Statements

Qualifying Statements

- This guidance represents the view of the National Institute for Health and Care Excellence (NICE), which was arrived at after careful consideration of the evidence available. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. However, the guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer, and informed by the summaries of product characteristics of any drugs.
- Implementation of this guidance is the responsibility of local commissioners and/or providers. Commissioners and providers are reminded that it is their responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties.
- The guideline will assume that prescribers will use a drug's summary of product characteristics to inform decisions made with individual patients.
- This guideline recommends some drugs for indications for which they do not have a UK marketing authorisation at the date of publication, if there is good evidence to support that use. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. The patient (or those with authority to give consent on their behalf) should provide informed consent, which should be documented. See the General Medical Council's Good practice in prescribing and managing medicines and devices for further information. Where recommendations have been made for the use of drugs outside their licensed indications ('off-label use'), these drugs are marked with a footnote in the recommendations.
- Patients and healthcare professionals have rights and responsibilities as set out in the National Health Service (NHS) Constitution for England – all NICE guidance is written to reflect these. Treatment and care should take into account individual needs and preferences. Patients should have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals. If someone does not have the capacity to make decisions, healthcare professionals should follow the Department of Health's advice on consent and the code of practice that accompanies the Mental Capacity Act and the supplementary code of practice on deprivation of liberty safeguards. In Wales, healthcare professionals should follow advice on consent from the Welsh Government.
- NICE has produced guidance on the components of good patient experience in adult NHS services. All healthcare professionals should follow the recommendations in Patient experience in adult NHS services.
- For all recommendations, NICE expects that there is discussion with the patient about the risks and benefits of the interventions, and their values and preferences. This discussion aims to help them to reach a fully informed decision (see also Patient-centred care).

Implementation of the Guideline

Description of Implementation Strategy

Implementation tools and resources to help practitioners put the guideline into practice are available on the National Institute for Health and Care Excellence (NICE) Web site ; see also the "Availability of Companion Documents" field.

Key Priorities for Implementation

The following recommendations have been identified as priorities for implementation.

Awareness of Clinical Features of Idiopathic Pulmonary Fibrosis

- Be aware of idiopathic pulmonary fibrosis when assessing a patient with the clinical features listed below and when considering requesting a chest X-ray or referring to a specialist:
  - Age over 45 years
  - Persistent breathlessness on exertion
  - Persistent cough
  - Bilateral inspiratory crackles when listening to the chest
  - Clubbing of the fingers
  - Normal spirometry or impaired spirometry usually with a restrictive pattern but sometimes with an obstructive pattern

Diagnosis

- Diagnose idiopathic pulmonary fibrosis only with the consensus of the multidisciplinary team (listed in table 1 below), based on:
  - The clinical features, lung function and radiological findings
  - Pathology when indicated

Table 1: Minimum Composition of Multidisciplinary Team Involved in Diagnosing Idiopathic Pulmonary Fibrosis

Stage of Diagnostic Care Pathway	Multidisciplinary Team Composition (All Healthcare Professionals Should Have Expertise in Interstitial Lung Disease)
After clinical evaluation, baseline lung function and CT	Consultant respiratory physician Consultant radiologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering performing bronchoalveolar lavage, and/or transbronchial biopsy or surgical lung biopsy Only some patients will have bronchoalveolar lavage or transbronchial biopsy but they may be being considered for surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Thoracic surgeon as appropriate Interstitial lung disease specialist nurse Multidisciplinary team coordinator
When considering results of bronchoalveolar lavage, transbronchial biopsy or surgical lung biopsy	Consultant respiratory physician Consultant radiologist Consultant histopathologist Interstitial lung disease specialist nurse Multidisciplinary team coordinator
See Chapter 6.5 (Multidisciplinary Team) in the full guideline document for more information on the expertise of the multidisciplinary team.	

Information and Support

- The consultant respiratory physician or interstitial lung disease specialist nurse should provide accurate and clear information (verbal and written) to people with idiopathic pulmonary fibrosis, and their families and carers with the person's consent. This should include information about investigations, diagnosis and management.
- An interstitial lung disease specialist nurse should be available at all stages of the care pathway to provide information and support to people with idiopathic pulmonary fibrosis and their families and carers with the person's consent.

Pulmonary Rehabilitation

- Assess people with idiopathic pulmonary fibrosis for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6-minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality-of-life assessment.

Best Supportive Care

- Offer best supportive care to people with idiopathic pulmonary fibrosis from the point of diagnosis. Best supportive care should be tailored to disease severity, rate of progression, and the person's preference, and should include if appropriate:
  - Information and support
  - Symptom relief
  - Management of comorbidities
  - Withdrawal of therapies suspected to be ineffective or causing harm
  - End of life care
- If the person is breathless on exertion consider assessment for:
  - The causes of breathlessness and degree of hypoxia and
  - Ambulatory oxygen therapy and long-term oxygen therapy and/or
  - Pulmonary rehabilitation

Disease-Modifying Pharmacological Interventions

- For guidance on pirfenidone for the management of idiopathic pulmonary fibrosis, refer to Pirfenidone for the treatment of idiopathic pulmonary fibrosis (NICE technology appraisal guidance 282).
- Do not use any of the drugs below, either alone or in combination, to modify disease progression in idiopathic pulmonary fibrosis:
  - Ambrisentan
  - Azathioprine
  - Bosentan
  - Co-trimoxazole
  - Mycophenolate mofetil
  - Prednisolone
  - Sildenafil
  - Warfarin

Lung Transplantation

- Refer people with idiopathic pulmonary fibrosis for lung transplantation assessment if they wish to explore lung transplantation and if there are no absolute contraindications. Ask the transplant centre for an initial response within 4 weeks.

Review and Follow-Up

- In follow-up appointments for people with idiopathic pulmonary fibrosis:
  - Assess lung function
  - Assess for oxygen therapy
  - Assess for pulmonary rehabilitation
  - Offer smoking cessation advice, in line with the NICE guideline Smoking cessation services in primary care, pharmacies, local authorities and workplaces, particularly for manual working groups, pregnant women and hard to reach communities (NICE public health guidance 10)
  - Identify exacerbations and previous respiratory hospital admissions
  - Consider referral for assessment for lung transplantation in people who do not have absolute contraindications
  - Consider psychosocial needs and referral to relevant services as appropriate

- Consider referral to palliative care services
- Assess for comorbidities (which may include anxiety, bronchiectasis, depression, diabetes, dyspepsia, ischaemic heart disease, lung cancer and pulmonary hypertension)

Implementation Tools

Audit Criteria/Indicators

Clinical Algorithm

Foreign Language Translations

Mobile Device Resources

Patient Resources

Resources

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

End of Life Care

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

National Clinical Guideline Centre. Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. 32 p. (Clinical guideline; no. 163).

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

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Guideline Developer(s)

National Clinical Guideline Centre - National Government Agency [Non-U.S.]

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National Institute for Health and Care Excellence (NICE)

Guideline Committee

Guideline Development Group (GDG)

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Financial Disclosures/Conflicts of Interest

At the start of the guideline development process all Guideline Development Group (GDG) members declared interests including consultancies, fee-paid work, share-holdings, fellowships and support from the healthcare industry. At all subsequent GDG meetings, members declared arising conflicts of interest, which were also recorded.

Members were either required to withdraw completely or for part of the discussion if their declared interest made it appropriate. The details of declared interests and the actions taken are shown in Appendix B of the full version of the guideline (see the "Availability of Companion Documents" field).

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the National Institute for Health and Care Excellence (NICE) Web site .

Availability of Companion Documents

The following are available:

- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Full guideline. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. 307 p. (Clinical guideline; no. 163). Electronic copies: Available in Portable Document Format (PDF) from the National Institute for Health and Care Excellence (NICE) Web site .
- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Appendices. London (UK):

National Institute for Health and Care Excellence (NICE); 2013 Jun. 493 p. (Clinical guideline; no. 163). Electronic copies: Available in PDF from the NICE Web site.

- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Baseline assessment tool. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. (Clinical guideline; no. 163). Electronic copies: Available from the NICE Web site.
- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Clinical audit tool. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. (Clinical guideline; no. 163). Electronic copies: Available from the NICE Web site.
- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Costing report. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. (Clinical guideline; no. 163). Electronic copies: Available from the NICE Web site.
- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Costing template. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. 32 p. (Clinical guideline; no. 163). Electronic copies: Available in PDF from the NICE Web site.
- Idiopathic pulmonary fibrosis. The diagnosis and management of suspected idiopathic pulmonary fibrosis. Podcast. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. (Clinical guideline; no. 163). Electronic copies: Available from the NICE Web site.
- Idiopathic pulmonary fibrosis: overview. NICE pathway. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. (Clinical guideline; no. 163). Electronic copies: Available from the NICE Web site.
- The guidelines manual 2009. London (UK): National Institute for Health and Care Excellence (NICE); 2009 Jan. Electronic copies: Available from the NICE Web site.
- The guidelines manual 2012. London (UK): National Institute for Health and Care Excellence (NICE); 2012 Nov. Electronic copies: Available in (PDF) from the NICE Web site.

Patient Resources

The following is available:

- Idiopathic pulmonary fibrosis. Information for the public. London (UK): National Institute for Health and Care Excellence (NICE); 2013 Jun. (Clinical guideline; no. 163). Electronic copies: Available from the National Institute for Health and Care Excellence (NICE) Web site. Also available for download as a Kindle or EPUB ebook from the NICE Web site. Also available in Welsh from the NICE Web site.

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status

This summary was completed by ECRI Institute on October 17, 2013. This summary was updated by ECRI Institute on June 2, 2016 following the U.S. Food and Drug Administration advisory on Opioid pain medicines.

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